Leptomeningeal dissemination of spinal pilocytic astrocytoma

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TITLE OF CASE

Leptomeningeal dissemination of Spinal Pilocytic Astrocytoma: A rare entity.

DESCRIPTION ***Up to 250 words summarising the importance of the image(s)***

A currently 12 year old boy presented at the age of 8-months with sleepiness, irritability and a tense fontanelle with upgaze palsy on a background of a 2 month history of vomiting, difficulty feeding and weight loss. Acute communicating hydrocephalus was diagnosed using ultrasonography and CT and treated with VP shunt insertion. Further investigation with MR imaging (2006) found a thoracic intramedullary mass with further intracranial and spinal leptomeningeal dissemination (Figures 1 and 2). A biopsy of the thoracic intramedullary mass showed pilocytic astrocytoma (PA) (Figure 3).

He received intensive 85 weeks of chemotherapy (2006-2008) with low grade glioma protocol (SIOP-LGG 2004), including Vincristine (1.125mg/m2 to 1.5mg/m2), carboplatin (412mg/m2-550mg/m2) and etoposide (100mg/m2). He responded well with almost complete resolution of metastatic disease (figure 4). There was no progression for several years till December 2016, when he developed a suprasellar/prepontine cystic area/loculated fluid with small eccentric enhancing nodule (figure 5); On further follow up, the cyst has become more well defined in 2017 and 2018 scans with slight prominence of nodule (figure 6). The nodular enhancement in the spinal intramedullary lesion has also become slightly more prominent since june 2017 (Figure 6) with otherwise overall stable appearances, there has been however no change in neurological status. He is currently neurologically and developmentally well. His main problem has been progressive thoracic and lumbar scoliosis secondary to thoracic tumour; stabilised with bracing since 2011. Due to concerns of destabilisation, and following pubertal maturation, the patient underwent elective posterior spinal fusion T2-4 with instrumentation and bone graft. He underwent posterior spinal fusion in 2017. He remains well, 10 years post treatment, with ongoing disease surveillance.

PA is the most common paediatric brain tumour accounting for ~20% of paediatric brain tumours.[1] PA usually arise in the posterior fossa, hypothalamic region or optic chiasm (often associated with Neurofibromatosis-1), occasionally in the cerebral hemispheres, and extremely rarely, in the spinal cord. Almost all are associated with single abnormalities of the mitogen-activating-protein-kinase (MAPK) pathway. While PA are Grade 1 tumours, they occasionally can exhibit more aggressive behaviour such as malignant transformation, recurrence or leptomeningeal dissemination (LD).[2,3] LD in PA is a rare occurrence, with less than 100 reported cases in the literature and even more rarely occurring with a spinal primary with only 4 reported cases.[2,4] Around half of cases of LD present at diagnosis.[1] It is not known, why LD, particularly with a spinal primary is such a rare occurrence. There is a possibility that it represents a distinct entity or has a specific phenotype, however, this will need further analysis. While LD can present with hydrocephalus, many are identified incidentally.[4] Since the number of known LD are very low, there are no known factors

described to suggest its occurrence, however, any change in neurological status and any clinical suspicion of raised intracranial pressure should prompt further imaging to exclude its occurrence. PA is primarily treated with surgery, and occasionally, with radiotherapy and/or chemotherapy, while targeted therapies hold promise. Prognosis is excellent; with 10-year survival >90%, falling to <50% for patients with LD.[4] As this case demonstrates however that despite leptomeningeal dissemination, these patients can still have a relatively long and indolent course. This case report is intended to serve as a reminder of this rare and often chronic condition.

LEARNING POINTS/TAKE HOME MESSAGE ***2 to 3 bullet points – this is a required field***

- **Pilocytic astrocytomas, while Grade 1 tumours, can rarely metastasize with leptomeningeal dissemination.**
- **Leptomeningeal dissemination can present incidentally, either at diagnosis or later, but can be symptomatic and may present as hydrocephalus.**
- **Despite early leptomeningeal dissemination, the patients can have a long and indolent course with a relatively stable disease over long periods.**

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FIGURE/VIDEO CAPTIONS ***figures should NOT be embedded in this document***

Figure 1: Initial MRI Whole spine (2006). (a) T2 sagittal, (b) T1 sagittal and (c) Post contrast T1 sagittal. Black arrows in (a) and (b) show upper thoracic intramedullary mass. This white arrow in (c) shows no significant enhancement. Thick white arrows (c) shows leptomeningeal enhancement.

Figure 2: Initial MRI Brain (2006). (a) T1 sagittal without contrast. (b) T1 sagittal with contrast. (c-f). T1 axial post contrast images. White arrows in b-f show several areas of leptomeningeal dissemination.

Figure 3: Histopathology shows solid and less solid areas with Rosenthal fibres and elongated cell nuclei with long fibrillary processes consistent with Pilocytic astrocytoma.

Figure 4. Post chemotherapy scan (2008). (a) T2 sagittal of spine. (b) T1 post contrast sagittal of spine. (c,d), T1 post contrast axials brain. (a,b) show stable

appearances of intramedullary mass and resolution of leptomeningeal dissemination. (c,d) show resolution of intracranial leptomeningeal dissemination.

Figure 5. Follow up scan (2016). (a,b) T2 and T1 sagittal post contrast images of spine. (c,d,e) T1 post contrast axial, sagittal and coronal images of brain. (a,b) show stable appearance of intramedullary mass (white arrows). Thick white arrow in (d) shows a CSF/fluid loculation in suprasellar region with draping of optic tract over it (broken arrow). Thin white arrows in (c,e) show small eccentric enhancing nodule on left side.

Figure 6. Follow up scan (2017). (a,b) T2 and T1 sagittal post contrast images of spine. (c,d) T1 post contrast axial and sagittal images of brain. (a,b) show increasing scoliosis. White arrow in (b) show nodular enhancing lesion slightly more prominent than previous images. White arrow in (c) shows nodular enhancement in suprasellar resion. White arrow in (d) shows more well defined fluid/loculated component.

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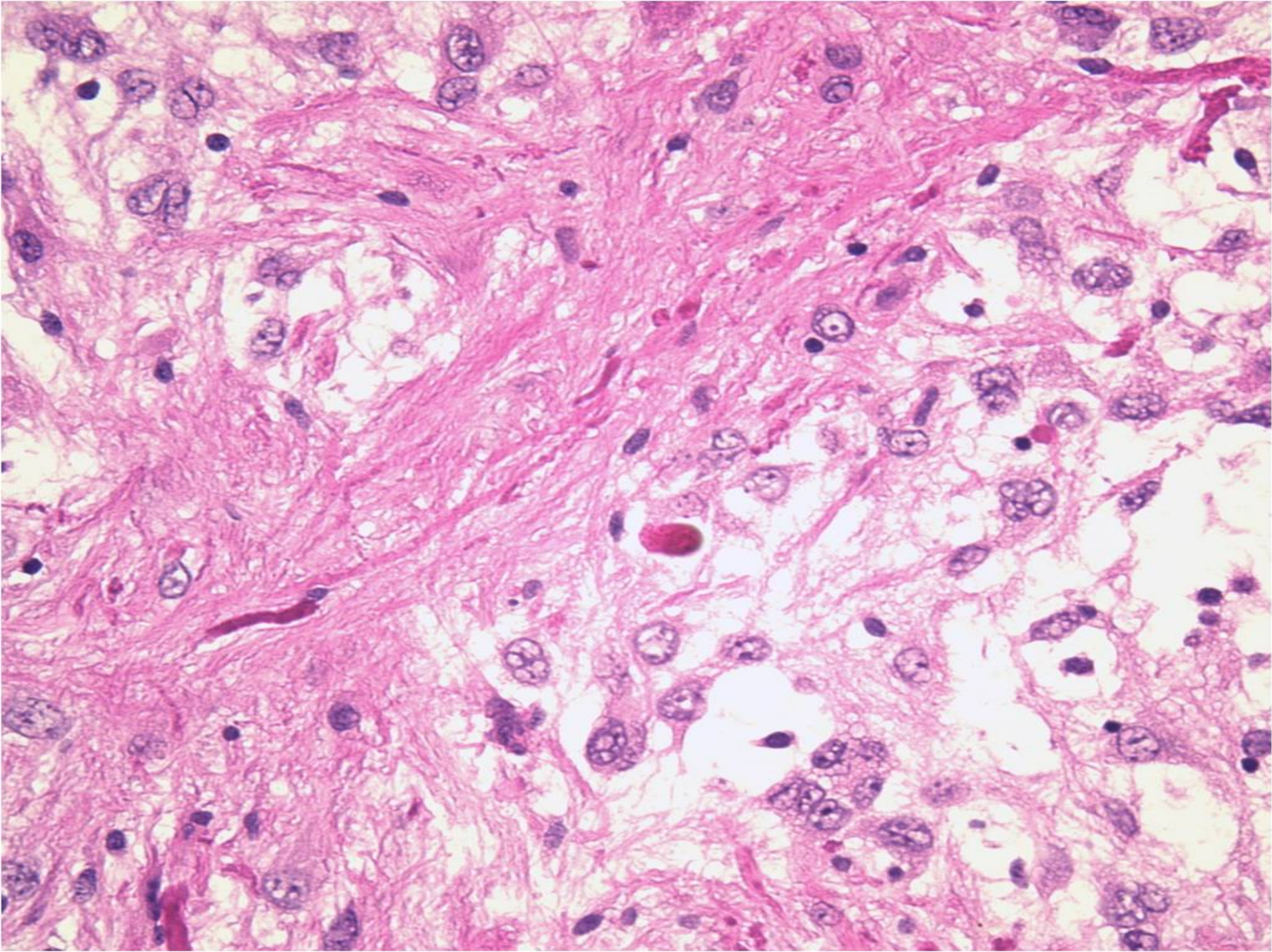

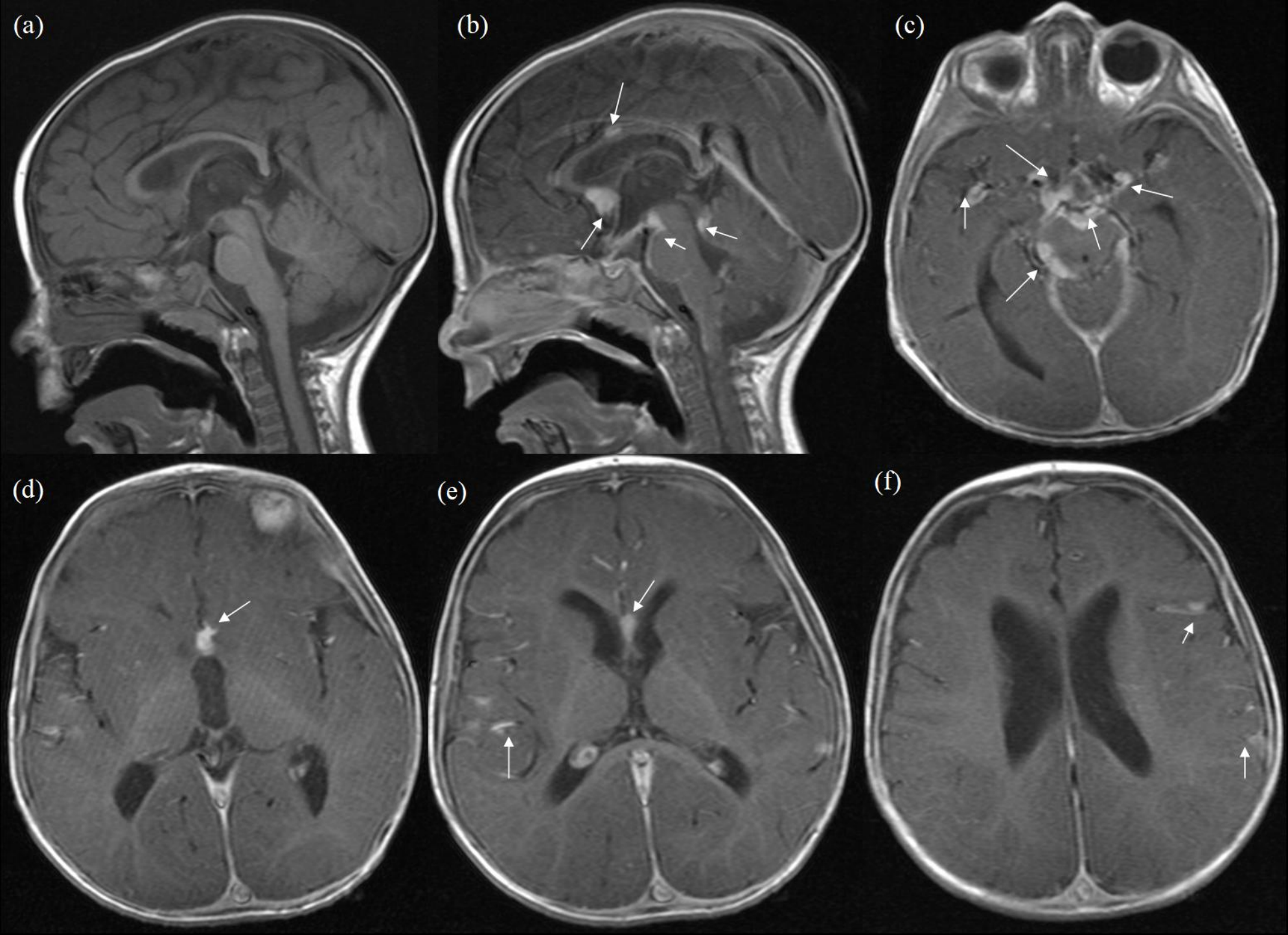
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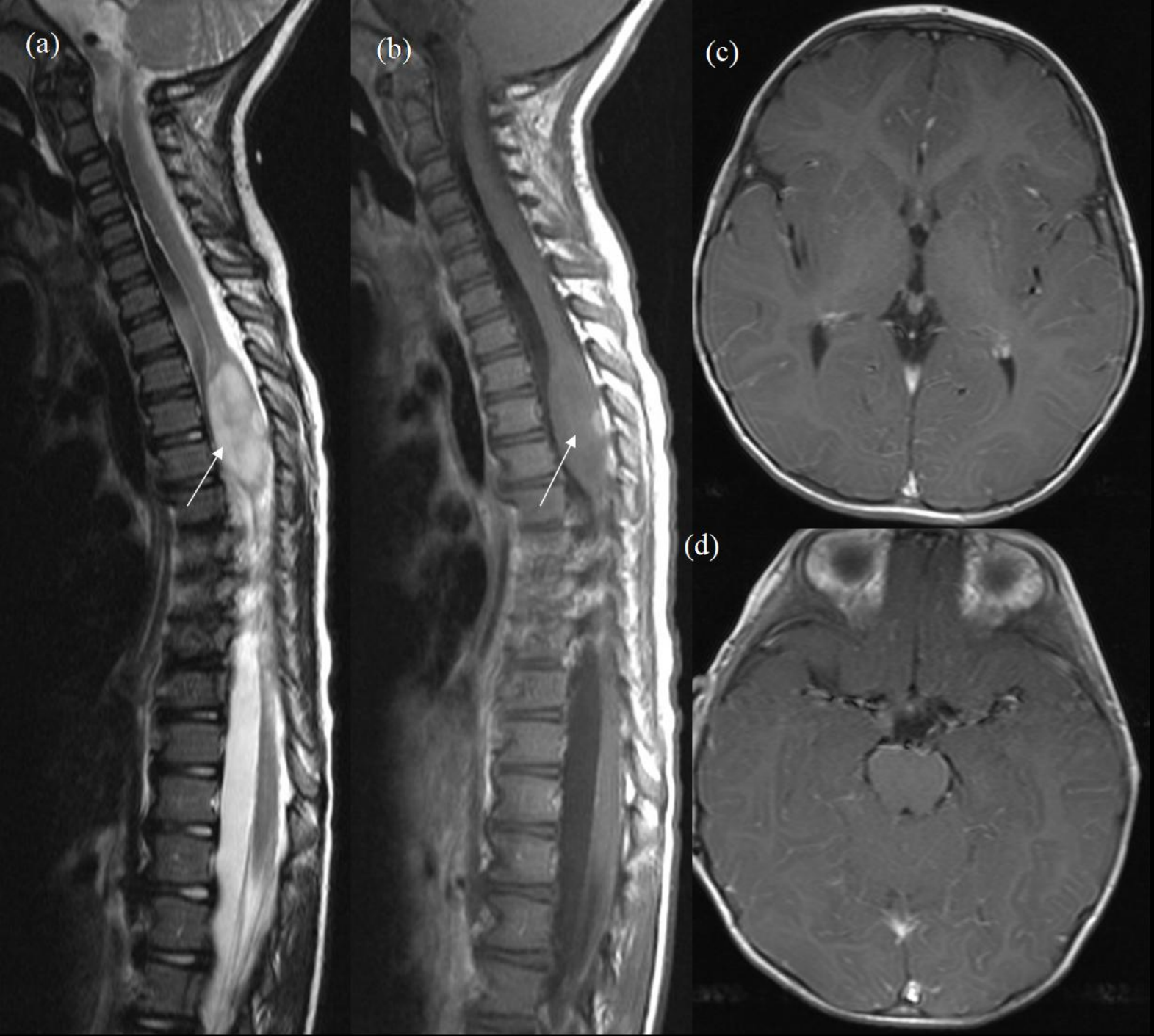
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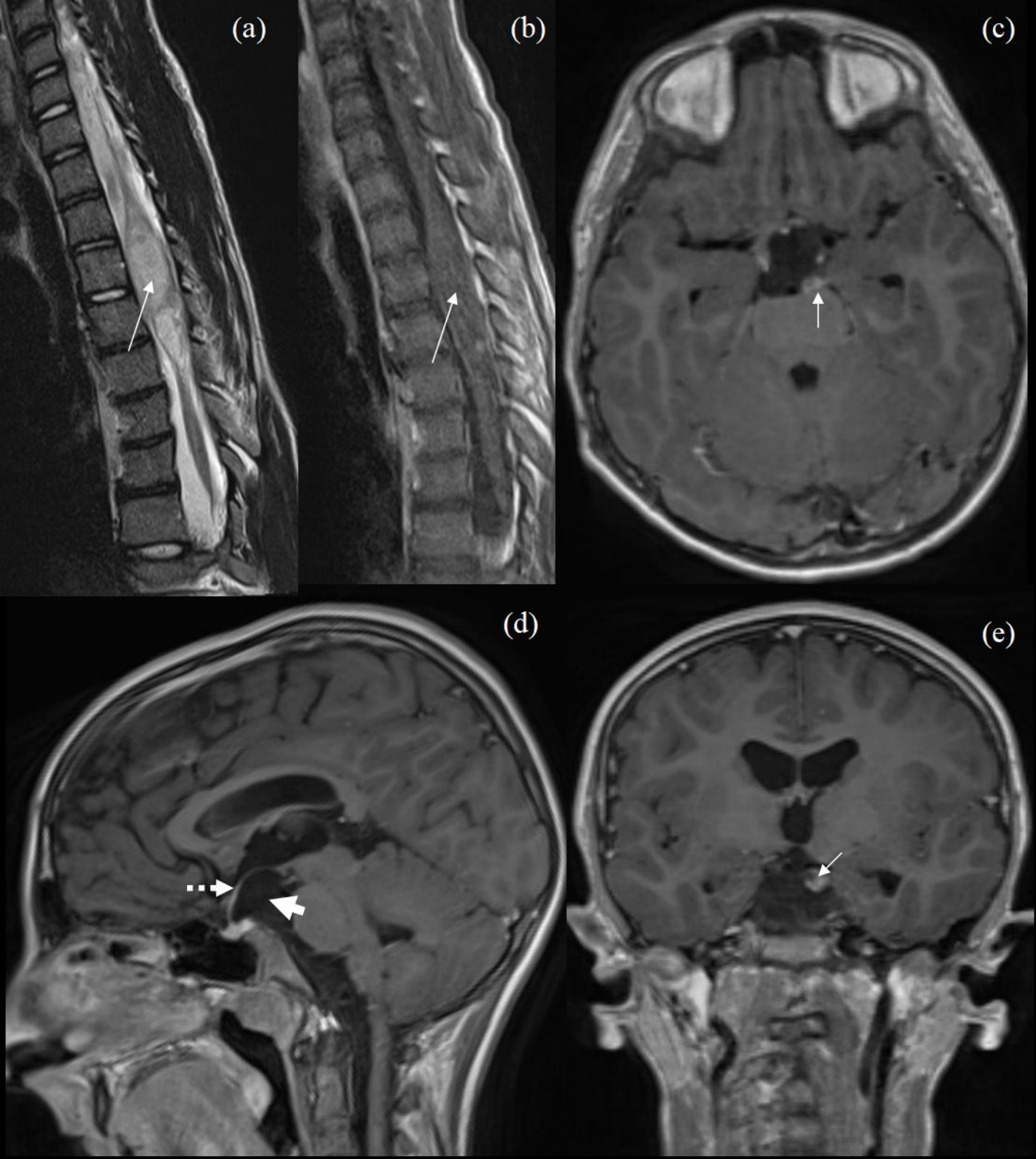
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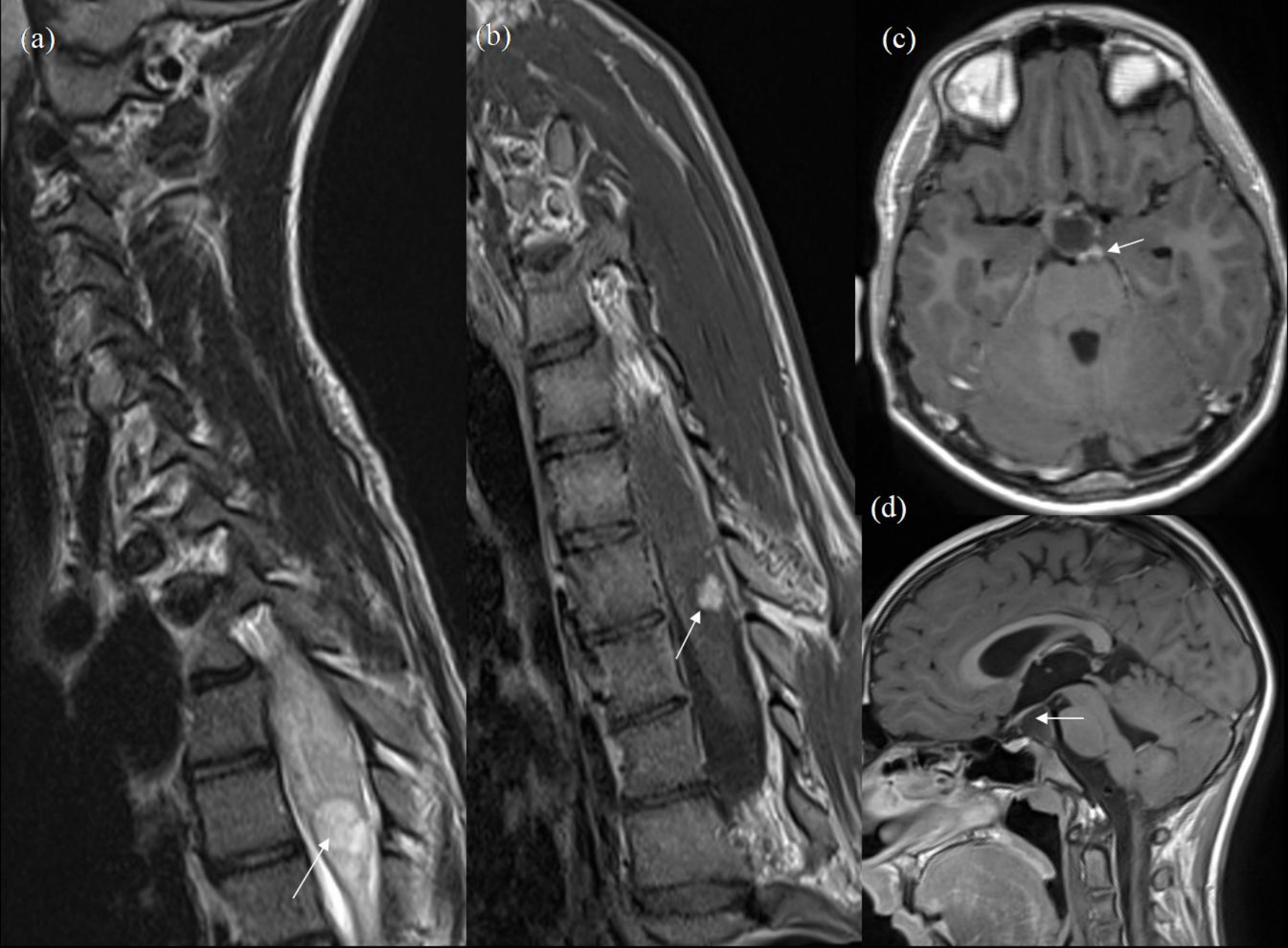
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